

SURGICAL ASPECTS OF TABES DORSALIS*†

BY

ROBERT LEES

Manchester

Tabes dorsalis is a convenient label for a complex group of neurological symptoms and signs caused by syphilis. The main lesions are in the posterior columns of the spinal cord, and the symptoms are therefore sensory. But there are also lesions affecting the autonomic nervous system, whose effects are on the viscera, the pupils of the eyes, and the limbs. In addition, this chronic, painful, and disabling disease, associated with syphilis, has a profound effect on the patient's mind.

There are few surgeons who have not been called upon to make the difficult decision whether operation be required in a patient who has symptoms of an abdominal disaster, and at the same time a history and signs of nervous disease. There are few physicians who have not diagnosed tabes dorsalis, and have pointed with scorn at the abdominal scars of useless operations. It is not my purpose to dwell on this contentious field but to discuss certain features of the disease which may interest both surgeon and physician alike. Before leaving it, however, I would point out that in a review of 1,000 cases of tabes by Nuzum 97 useless operations were recorded (cited by Stokes and others, 1944, p. 1013).

Surgical Diagnosis	No. of Operations
Gastric ulcer	19
Gall stones or cholecystitis	19
Appendicitis	18
Salpingitis	13
Exploratory laparotomy	9
Renal calculi	7
Post operative adhesions	7
Others	5
Total	97

Morbid Anatomy.—The pathological changes found in the spinal column are well known, but their causation is not understood, and there is much work to be done on the elucidation of the problems of this disease. It is usually assumed that the primary attack is on the posterior nerve root

fibres as they pass from the ganglia through the meninges to the cord. The posterior nerve roots of the affected spinal segments may present degenerative changes and there is frequently a great increase of glial fibres at the entrance zone of these fibres. Fibrous tissue is increased around the dorsal root ganglia, and the meninges are often thickened, especially over the posterior part of the cord. Several hypotheses have been put forward, but there is no proof or satisfactory explanation for the very selective action on the posterior roots and the ascending degeneration of the posterior columns of Goll and Burdach. The pupil changes are probably due to gliosis around the aqueduct of Sylvius at the level of the 3rd nucleus. The optic nerve shows degenerative changes, primary optic atrophy occurring in about 10 per cent. of tabetics. The 5th, 8th, and 9th cranial nerves, may also suffer degenerative changes.

There is very little evidence of pathological changes in the ganglia of the paravertebral ganglia of the autonomic nervous system and the rami communicantes, but it is suggested that such changes may be the cause of many of the symptoms of tabes dorsalis, and may afford the explanation for so-called "burnt-out" tabes or the "formes frustes" of Fournier, in which optic atrophy or visceral pains may be the main evidence of the disease.

It has been suggested that vitamin deficiencies, particularly of the B complex, may be responsible for the selective action of syphilis on the sensory and autonomic nerve fibres. But the evidence is not convincing, and treatment with vitamin concentrates seldom produces amelioration of the disease.

Serological Changes.—The blood gives a positive serum test for syphilis in about 80 per cent. of cases. The changes in the cerebrospinal fluid are well known: slight pleocytosis, increase of globulin and of total protein, a positive Wassermann reaction in about 75–80 per cent. of cases, and changes

* Received for publication April 30, 1952.

† Address given at the Royal College of Surgeons of England in 1951.

in the Lange gold sol curve, most marked in the second zone.

Morbid Changes in Other Organs.—As tabes dorsalis usually occurs 15 or more years after the primary infection with syphilis, it affects middle-aged persons, and other effects of syphilis, particularly in the heart and aorta, may also be present. It is not uncommon for the cardiovascular disease, though silent, to be of much graver significance to the patient. In late cases, and especially neglected cases, such as fill "chronic" wards, the patients have severe urinary tract infection, often with pyelonephritis and impending uraemia. Taboparesis is not uncommon, but as a rule does not have the rapid downward course of general paresis.

Symptoms of Tabes Dorsalis

The clinical picture is very variable, but a composite picture of the disease is clearly shown in the following tabulation (Stokes and others, 1944, p. 1011):

Important Symptoms and Signs of Tabes Dorsalis

<i>Symptoms</i>	<i>Per cent.</i>
Lightning pains	73-88
Visual symptoms	44
Difficulty in urination—Trouble in starting, dribbling afterward, nocturnal frequency and "bed-wetting"	43-68
Paresthesias (numbness, prickling, girdle sensation, etc.)	40-58
Ataxia	37-87
Strabismus	12
Diplopia	11
Girdle sense	10-31
Visceral crises (gastric, rectal, laryngeal) ..	10-22
Loss of libido and potentia	6-25
Failing vision	6
Ptosis	5-23
Vertigo	4-13
Deafness	1-4
<i>Signs</i>	
Argyll Robertson pupils	80
Reduced or absent lower cord reflexes ..	70-90
Romberg sign	43-96
Sensory disturbance and dissociation ..	49-58
Diminished pain	
Diminished vibration sense (bone fork conduction)	
Diminished sense of motion and position	
"Cord" (atonic) bladder	43
Trophic changes—Charcot joints, malum perforans	12-19
Optic atrophy	6-9
Miscellaneous, including muscle atrophy, tabetic clubfoot, wrist and foot drop, hyperextensibility of joints (decreased muscle tone) mentioned by various authors	

It is emphasized that it is the early symptoms, and the incomplete forms with few signs, which present the difficult surgical problems. The diagnosis

can and should be made on grounds of history and signs and without reference to the results of blood and cerebrospinal fluid examinations which are of more value in prognosis than in diagnosis.

Pain is almost always an accompaniment of tabes. The pain may set in early, it may precede the development of the disease picture by months or years, or it may accompany it throughout its entire course. The pain may be mild or severe, sharp, boring, shooting, stabbing, cutting, lightning-like, or compressing like a girdle. It may be like a mild rheumatism, or pass for sciatica. Frequently it is paroxysmal and lasts for a few seconds to a few hours. But the site of the pain depends on the nerve roots involved. If the lumbosacral roots are involved there are pains in the legs; if the lower dorsal roots are affected the pains are in the abdomen and it is in the latter cases that early tabetics are operated on for appendicitis, gall stones, or salpingitis. Besides the pains the patient may complain of dysaesthesias or peculiar disagreeable sensations in various radicular zones.

If the inflammatory processes affect the sympathetic rami, then visceral pains result, and these are described as "crises". According to the roots and rami involved there may result rectal, vesical, or sexual crises, or gastric, or laryngeal crises. The gastric type is much the most common. The crises are paroxysmal in nature, come on suddenly, last a few hours, days, or weeks, and disappear suddenly, only to recur again days or weeks later.

Three types of gastric crises are described:

- (a) the attack of pain without vomiting;
- (b) the attack of vomiting without pain;
- (c) the most common type with both pain and vomiting.

Exhaustion, emotional stress, cold, damp weather, acute infections, and treatment shocks including lumbar puncture, may serve as precipitating incidents, but often the attack comes from a clear sky. In the splanchnic type of crisis there is pain and hyperalgesia of the abdominal wall without true deep pain or rigidity. Intense nausea may be the main symptom. The vomiting may be intense, continuous, and intractable, the stomach contents being ejected with violence, and this may occur with little nausea.

Crises in the abdomen have been classified as:

- (i) sympathetic, with marked pain, vomiting, and hyperalgesia over the abdomen;
- (ii) vagal with little pain, but marked nausea and vomiting, with pain in the region of the larynx, ear, or heart;
- (iii) phrenic crises with abdominal pain, hiccough, and pain over one shoulder.

Radiologically it has been proved that during crises the stomach is in violent contractions, and this may be shared by the entire gut.

In some patients spastic constipation periodically alternates with diarrhoea.

In early tabes there may be an imperious demand for immediate evacuation of the bladder. This urgent desire to micturate cannot be satisfied, and by straining or posture the patient may succeed in voiding a small amount of blood-stained urine. This may be accompanied by a sudden intense shooting pain along the penis and into the testes.

Organic disease of abdominal organs may co-exist with tabes dorsalis. Recently I made a study of a group of tabetics to ascertain if there was an abnormal incidence of peptic ulcer, but I could not find evidence to suggest that the incidence of peptic ulcer in tabetics differed from that in a comparable group of non-syphilitic hospital out-patients.

It may be difficult to decide if a patient's symptoms are attributable to tabes dorsalis, or to disease of the abdominal viscera, such as peptic ulcer or gall bladder disease, or to functional nervous disease. I well remember a miserable tabetic man, who complained week after week of rather indefinite and mild digestive symptoms. I fear that we tended to dismiss this as the habitual complaints of a querulous and introspective patient until he was admitted to hospital after a severe haematemesis from which he ultimately died. Necropsy revealed a large, deep, and chronic gastric ulcer. Since then I have always subjected tabetics with digestive symptoms to radiological investigation.

Treatment of Gastric Crises

These are self-limiting, but the patient may become dehydrated and exhausted, and lose weight rapidly in a prolonged bout; he fears the recurrence of crises, and demands relief.

Atropine in large doses may alleviate pain; it is wise to avoid morphine and all opiates for the danger of drug addiction is great. Paravertebral injection of novocaine, diathermy, or deep x ray in the lower dorsal region, may give temporary relief. Also advocated are the injection of 5-10 units insulin, or intravenous infusion of glucose saline.

It may be worth while to try the effect of hexamethonium compounds, which block the synapses in the ganglia of the autonomic nervous system. Between attacks the patient must have rest, nourishing food to make up loss of weight, and mental reassurance.

Specific therapy of syphilis, including penicillin, is generally disappointing, though the frequency

and severity of bouts tend to be reduced by prolonged treatment.

Operative treatment, including section of a group of posterior nerve roots and chordotomy, is generally a last desperate remedy, fraught with danger and unlikely to succeed.

The Bladder

Stokes and others (1944), in a collation of 1,500 cases, report difficulty in urination in 43-68 per cent., and cord (atonic) bladder in 43 per cent. The healthy adult is normally unconscious of the rhythmic contraction of the bladder as it fills until the volume of urine stimulates such powerful contractions that the individual is conscious of distension, pain, and a desire to void urine. By exercise of voluntary control, which is an acquired faculty, the reflex contractions can be resisted for a time until social conditions permit urination. The urgency of the call to urinate is also conditioned by the state of the emotions. Fear may produce urgency or even incontinence; pre-occupation with other interests may prevent the bladder stimuli reaching consciousness.

Once urination has begun it tends reflexly to continue till the bladder is empty. An adult can interrupt the act, but perhaps not always successfully. But in the tabetic the sensory impulses from the bladder become fewer, and feebler, and delayed. The bladder must contain a greater volume of urine before the impulse to void becomes perceptible. When the urine flows the tabetic may not experience the sensation of urination and the desire to micturate is readily satisfied, before the bladder is empty. The tabetic therefore urinates more and more seldom, and he less and less completely empties his bladder. The bladder becomes progressively more distended, the residual urine increases, and infection is common, and this leads to severe intractable genito-urinary infection which is not infrequently the cause of early death. The tabetic may have no conscious desire to micturate, but may induce emptying by abdominal pressure, straining, and other devices, in order to avoid the miseries consequent on incontinent dribbling from overflow retention.

Cystoscopic Appearances.—The bladder has an excessive capacity, the walls are trabeculated, often atrophic, and there is often evidence of chronic infection. The appearances are those described as "cord" bladder.

Management of Bladder in Tabetics.—

(a) Prevent habitual over-distension by teaching the patient to void frequently and completely.

(b) Minimize risk of introduced infection by ensuring strict aseptic ritual, should the tabetic patient have to have catheterization, bladder lavage, etc.

(c) Tabetics with chronic distension of bladder with residual urine, should be handled in much the same way as cases of chronic or acute distension of bladder associated with prostatic disease.

(d) The following emergency measures are advocated :

(i) bladder drill to induce spontaneous contraction. This may be assisted by injections of choline derivatives—e.g. carbachol 1 cc. ;

(ii) aseptic catheterization twice daily, followed by irrigation through the catheter and finally per urethram using 1/10000 silver nitrate lotion. This should be regarded as a temporary measure and voluntary control secured as much as possible ;

(iii) with the above, appropriate chemotherapy with penicillin or other antibiotic, sulphonamide, or other appropriate antiseptic to control the infecting organisms.

Many tabetics who have reached the stage of overdistension, incontinence, and chronic infection can regain functional control of their urination, and when the danger of pyelonephritis and uraemia is removed, their lives are greatly prolonged. At the same time the patient's miseries are relieved and he becomes a more popular citizen, able to resume more active social life.

Arthropathies

In a small percentage (? 5 per cent.) of tabetics, lesions of the large joints appear which are ascribed to trophic changes consequent on disease affecting the trophic nerve fibres associated with the autonomic nervous system. The commonest are Charcot's joints, or tabetic arthropathies, but this category also includes pathological fractures and perforating ulcer of the foot. The knee, hip, ankle, and shoulder joints are most commonly affected. The vertebral column is occasionally involved, and the collapse of a vertebra may cause compression of the cord.

These are usually painless swellings of the joints with effusions and oedema, proliferation, destruction of the cartilages, capsule, and bony surfaces, and formation of new bone, fragments of which may be loose within the disorganized joint.

In the earliest stages the joint may be painful, but this is exceptional. Indeed it is astonishing how great destruction and distortion of the anatomical relations are possible without pain and with preservation of function. The cause of these arthropathies is assumed to be involvement of the sympathetic fibres. Another theory is that by loss of sense of

position and pain sensation, and hypotonia of the muscles, the joint is subjected to frequently repeated trauma, which eventually causes effusions and destruction of the articular surfaces. In particular, the onset of Charcot joint is often determined by trauma.

Among other arthropathies and atrophic effects may be classified perforating ulcer of the foot, which is usually associated with necrosis of the head of the first metatarsal bone, and a chronic infected but painless sinus.

Tabetic arthropathies require careful management :

(a) treatment of the tabetic process ;

(b) fitting by an orthopaedic surgeon of an appliance which will prevent weight bearing or similar trauma to the joint, but which may permit limited movement ;

(c) protection of other joints from excessive strain through the use of (b).

I have observed quite remarkable reductions in the size of the effusion, absorption of much of the excess bone formation, and preservation of a moderately useful joint after prolonged use of appropriate orthopaedic appliances, and I have also observed the development of arthropathies in the joints of the right side following the use of a walking calliper to protect the left knee joint.

Surgical excision of arthropathies is only occasionally advised. It is undertaken when the tabetic process has been well treated, when the arthropathy has not responded well to conservative measures, and where the patient is disabled by the arthropathy. There is a danger that sound arthrodesis may not be secured. Often the destructive process is so extensive that very wide excision of the joint is required, and it is difficult to secure a good functional result even with bone implants.

Contrary to expectation the results of conservative treatment of perforating ulcer of the foot are quite good. The patient should be fitted with a support so that the foot is relieved from weight bearing. Suitable dressings will control local infection of the sinus, sometimes after a preliminary removal of the surrounding necrotic horny material. It is rarely necessary to excise the head of the metatarsal. Eventually after 6-12 months, the sinus heals, but the area should be protected from pressure by wearing a foot pad or a calliper.

Ataxia

The surgeon is seldom concerned with this symptom, but I believe that a tabetic should hardly ever be allowed to become bed-ridden. Ataxia is invariably severe after a period of confinement to bed, and if surgery is needed the patient should be

made to get up as soon as possible. If the patient is unable to leave his bed for some time, the nurse or physiotherapist should see that he performs simple movements to maintain muscle tone and preserve muscle control.

Psychological Factors

These are of considerable importance and may dominate the picture. Every tabetic has a great anxiety or even fear which accentuates the symptoms and may determine the onset of visceral crises. Certainly fear aggravates visceral pains, ataxia, incontinence of urine, and sexual impotence. Many tabetics become depressed, hopeless, helpless, bed-ridden invalids. Many become confirmed addicts to sedatives and opiates. Some become worn out by ceaseless pain and misery, and overwhelmed by shame and remorse. Very few resort to suicide but to many death comes as a merciful release.

It is an axiom that a hopeful prognosis should be given in the management of tabes. If a reasonably early diagnosis is made the outlook for life is quite good and many live to a ripe old age. Relief from symptoms is usually considerable, though pains and crises may be resistant, and may defy the therapeutic resources of the doctor. Even the advanced cases can be given some hope of improvement, unless neglect of infection and gross degenerative changes render recovery impossible.

Summary

The surgical aspects of tabes dorsalis are :

- (a) the visceral crises, which may produce difficult diagnostic problems ;
- (b) urinary symptoms ;
- (c) arthropathies.

It is considered that most of these are due to involvement of the sympathetic and vagus fibres, especially in the ganglia and rami communicantes, associated with the posterior roots of the spinal nerves.

Specific treatment of syphilis and special treatment of symptoms is generally rewarded by a great measure of relief.

Prevention of urinary complications or treatment of established "cord" bladder is of life-saving importance to the tabetic.

Tabetics should be kept as mobile as possible to prevent severe ataxia. If ataxia has developed it is possible to re-educate the patient to a very great extent.

Arthropathies respond moderately well to conservative orthopaedic procedures ; it is occasionally necessary to undertake excision, arthrodesis, or bone transplantation.

REFERENCE

- Stokes, J. H., Beerman, H., and Ingraham, N. R. (1944). "Modern Clinical Syphilology", 3rd ed. Saunders, Philadelphia and London.